

372 Routine vitamin supplementation provides normal serum vitamin A but not vitamin E concentrations in CF patients

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Cystic fibrosis is a complex disease and leads to the deficiency of fat soluble vitamins. Therefore, patients routinely undergo supplementation.

The aim of the study was: (1) to assess serum concentrations of vitamin A and E, (2) to analyze the relationship between clinical expression of the disease and body resources of these vitamins.

Material and Methods: The study comprised 43 CF patients in whom serum vitamin A and E concentrations were determined (HPLC). The nutritional status was assessed on the basis of Z-scores for body height and weight. Exocrine pancreatic secretion (fecal elastase-1, ELISA) and lung function (FEV1) served as markers of CF clinical expression.

Results: The ranges of vitamin A and E concentrations (median; mean±SEM) were as follow: 133–872 ng/mL (351; 377±26) and undetectable–15.6 µg/mL (8.9; 8.5±0.7). In all but one CF patients vitamin A levels were normal. Undetectable and abnormal serum vitamin E levels were found in 17 (39.5%) and 35 (81.4%) patients, respectively. Significantly lower vitamin E concentrations ($p < 0.04$) were found in subjects with worse nutritional status. Similarly, vitamin E body resources correlated to fecal elastase-1 ($r = 0.406$, $p < 0.006$).

Conclusions: Routine vitamin supplementation with available in Poland water-soluble preparation provides normal serum vitamin A concentrations. However, it is ineffective in case of vitamin E. It seems that clinical expression of the disease plays some role for body resources of vitamin E and does not influence significantly vitamin A levels.

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374* Linoleic acid but not EPA and DHA correlates to prognostic markers in Scandinavian CF patients

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CF patients may have deranged lipid metabolism. We wanted to determine the pattern of fatty acids (FA) in serum phospholipids (PLs) of Scandinavian CF patients and correlate the pattern to nutritional status and prognostic parameters as a basis for further studies.

Patients: 710 CF patients (362 males), mean age 20.4 (median 18.0, range 0.3–65.9) yrs were investigated. 86% were pancreatic insufficient.

Methods: The FA pattern in s-PLs of each patient was analyzed with capillary gas chromatography. Linoleic (LA), arachidonic (AA), eicosapentaenoic (EPA) and docosahexaenoic (DHA) acid were correlated with the PUFA intake calculated from a 7 day dietary food record ($n = 343$), and with different clinical parameters.

Results: Mean (mole%) was for LA 16.0, AA 7.5, EPA 1.0, DHA 2.6. In Norway more patients showed higher levels of EPA and DHA compared to patients in Denmark and Sweden. Females had a higher DHA level than males (2.7 vs 2.5, $p < 0.002$). There were no other gender differences, nor any age differences. There was a positive correlation between LA and FEV1.0 (all ages), $p < 0.001$ and a negative correlation with HbA1c, $p < 0.00001$. There were no correlations between n-3 fatty acids (EPA and DHA) and IgG but a negative one with LA.

Conclusion: Generally PUFA levels were low showing different patterns. Unexpectedly, LA but not EPA and DHA correlated with FEV1 and HbA1c. This warrants further investigation.

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373 Vitamin D insufficiency in cystic fibrosis patients of central region of Russia

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Aims: to assess the vitamin D status of CF patients with different degree of exocrine pancreatic insufficiency taking pancreatic enzymes supplements and to compare it with vitamin D level in the reference group.

Methods: serum 25-hydroxyvitamin D (25OHD) level was examined in 24 patients with CF aged 2–22 years, related to faecal elastase-1 (FE-1) values, disease severity, serum calcium concentration and compared to the age- and gender-matched control with chronic lung diseases.

Results: All CF patients had low levels of 25OHD (< 40 ng/mL). 41.7% of the patients were vitamin D insufficient (< 20 ng/mL), in 16.6% children severely low vitamin D levels (< 10 ng/mL) were seen. In 16.6% control subjects 25OHD level was normal. In the 25OHD < 20 ng/mL and < 10 ng/mL groups were only 12.5% and 4.2% of the patients respectively. Mean values of 25OHD content during the dark season (16.28 ± 8.54 ; 22 ± 13.1) were significantly lower as compared with the bright season (27.9 ± 13.2 ; 34.52 ± 9.4 respectively) in both groups. 25OHD levels were decreased in CF patients during the whole year as compared with control subjects. Vitamin D insufficiency was associated with the age growth. No relationship with 25OHD content and gender, CF severity, genotype was found. The serum levels of calcium were lower than normal in 33.3% CF cases but there was no correlation with 25OHD concentration. The values of FE-1 were decreased in 83.3% CF patients, in 16.7% cases pancreatic activity was normal. No correlation between pancreatic function measured as FE-1 and 25OHD concentration, serum levels of calcium was noted.

Conclusion: Despite the adequate supplementation of pancreatic enzymes and absence of clinical symptoms of malabsorption serum 25OHD concentration remains low in children and adolescents with CF.

375* Clinical effect of long term oral DHA supplementation in cystic fibrosis patients

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Aim: The study objective was to evaluate whether the earlier observed changes in the fatty acid profile as result of DHA supplementation, could be maintained during longer study trials and to observe its effect on the clinical outcome of cystic fibrosis (CF) patients.

Method: A 1 year lasting double blind placebo controlled study was performed in $\Delta F508$ homozygous CF patients above the age of 6. Clinical data including pulmonary function and number of infections were collected. Blood for the determination of the fatty acid (FA) composition of serum phospholipid, vitamin E, liver enzymes, immunoglobulins, erythrocyte sedimentation rate and coagulation was drawn at the beginning and every 6 months after the start of the study.

Results: Seventeen patients were included, one dropped out. The treatment group was supplemented with an algal DHA-rich oil and the control group with sunflower seed oil. There was no difference between controls and treatment group for W/H%, caloric intake, FEV1% and FVC% at the start of the study and after 1 year of supplements. The phospholipid FA composition did not change in the control group. The treatment group had a significant increase in DHA and eicosapentaenoic acid (EPA) concentration. A concomitant decrease of dihomo-gammalinolenic acid, arachidonic acid, 22:5 n-6 and Mead acid was observed. The laboratory results showed no changes in vitamin E level, liver enzymes, albumin, erythrocyte sedimentation rate and IgG concentration in neither placebo nor intervention group.

Conclusion: Although DHA-rich oil shifted the serum phospholipid FAs to a less pro-inflammatory profile no conclusive clinical improvement could be observed so far.